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Sarcoidosis and skin manifestations

Aydan Yazıcı[1], Derya Hoşgün[2]

[1]Department of Dermatology, Aydın Atatürk State Hospital, Aydın, Turkiye
[2]Department of Chest Disease and Intensive Care Unit, Ankara Atatürk Sanatorium Training and Research Hospital, University of Health Sciences, Ankara, Turkiye

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Corresponding Author: Derya Hoşgün, deryahosgun@gmail.com

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ABSTRACT

Sarcoidosis is a multisystemic inflammatory disease of unknown etiology, affecting all organs and systems. Skin involvement is seen in one-third of cases. Cutaneous findings alone do not determine the prognosis of sarcoidosis, but they are useful in early diagnosis. There are specific and nonspecific cutaneous findings. The most common cutaneous findings are papules and plaques. This review summarizes the cutaneous manifestations of sarcoidosis.

Keywords: Inflammatory disease, sarcoidosis, skin manifestations

INTRODUCTION

Sarcoidosis is a systemic disease characterized by multisystemic inflammation of unknown cause. Histopathologically, it is characterized by noncaseating granulomas consisting of epithelioid histiocytes, giant cells, and lymphocytes composed of activated macrophages.[1] It is observed more frequently between 25 and 35 years of age and in women. Lung involvement is observed in 90% of cases. Involvement of the skin, gastrointestinal tract, lymph nodes, heart, and central nervous system is also observed.[2] Clinically, it can range from asymptomatic to acute, subacute, chronic, or multiple organ failure. In the lungs, spontaneous remission may be observed in pulmonary fibrosis.[3] Symptoms, clinical findings, and organ involvement vary according to race, age, and gender. The most common symptoms are malaise, fatigue, fever, night sweats, and weight loss.[2,3] After differential diagnosis, the diagnosis is evaluated with radiologic, clinical, and histopathologic findings. The first step in treatment should be decided according to sarcoidosis-related death, organ damage, and deterioration in quality of life.[4]

DERMATOLOGICAL FINDINGS

While skin involvement in sarcoidosis is observed in one-third of cases, it may pose a diagnostic challenge due to its different morphology. According to studies in the literature, lesions can be divided into specific and nonspecific lesions. Specific lesions are associated with chronic forms, while nonspecific lesions are associated with acute forms. Nonspecific lesions have a better prognosis. Papular, maculopapular, plaque, subcutaneous nodular, and ulcerated lesions are specific lesions. The most common nonspecific lesions are erythema nodosum, calcifications, and prurigo. The most common cutaneous findings are papules and plaques. In a study evaluating patients with cutaneous sarcoidosis, cutaneous lesions were the first finding in 74% of the patients. Cutaneous findings alone do not determine the prognosis of sarcoidosis. In general, early diagnosis is also beneficial.[5-9]

Maculopapular Sarcoidosis

It is the most common form. It is frequently localized on the face, especially around the eyes and nasolabial fold, but may also involve the occipital part of the neck, trunk, extremities, and mucous membranes. On examination, it is firm and has the consistency of apple jelly under pressure. It can be one cm small, red to brown, or purplish in color. Rosacea, sebaceous hyperplasia, Xanthoma, perioral dermatitis, tinea faciei, cheilitis granulomatosa, cutaneous Crohn's disease, granuloma faciale, lymphocytoma cutis (Lyme borreliosis), lupus vulgaris, lupus miliaris disseminatus faciei, lupus erythematosus, and secondary syphilis should be considered in differential diagnosis. Lesions may regress spontaneously or coalesce into plaque or annular lesions. Clinically, it is frequently detected at the onset of sarcoidosis.[5-8] Trauma areas predispose to maculopapular areas. In the literature, it has been found that maculopapular lesions mostly heal spontaneously or with treatment in less than 2 years without scarring.[9,10]

Plaque Lesions

On examination, they are symmetrical, bilateral, oval, red, and brown lesions with the consistency of apple jelly. They are seen on the face, extremities, and trunk. They are raised lesions measuring more than 5 mm. They tend to be thick, hard, and scaly. Lesion colors can vary from red to brown and from brown

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to yellow. Lichen planus, granuloma annulare, discoid eczema, syphilis, mycosis fungoides, tinea corporis, leishmaniasis, non-tuberculous mycobacteriosis, psoriasis, and cutaneous T-cell lymphoma should be considered in the differential diagnosis. Plaques tend to recur after treatment. The rate of healing with permanent scarring is higher than in papular sarcoidosis. It is associated with the chronic form of sarcoidosis.[5,6,8-10]

Scar Sarcoidosis

They are patchy, erythematous, or violet lesions that occur in areas of scar tissue due to previous trauma, surgical scars, vaccination, or herpes zoster, affecting all areas. They are often asymptomatic and associated with sarcoidosis exacerbations. Keloid or hypertrophic scars should be considered in the differential diagnosis. 29% of the studies have been found. There are cases associated with pulmonary or mediastinal involvement of sarcoidosis. Because tattoos can also cause sarcoidosis, patients should avoid getting them.[5,6,8-10]

Subcutaneous Nodular Lesions

It is also known as Darier-Roussy sarcoidosis. It is often localized in the extremities and can be seen in the eyes and face. There is no systemic involvement and it is not a sensitive lesion. They are hard, oval, multiple localized skin-colored lesions. They are 0.5-2 cm in size. They are more common in women in the fourth decade. They are mostly asymptomatic. Granuloma annulare, rheumatoid nodules, xanthomas, and lipomas should be considered in the differential diagnosis.[5,6,8-10]

Ulcerated Lesions

They are often early manifestations of systemic sarcoidosis, but they can also develop over previous sarcoidosis skin involvement. Mucous membranes and the scalp are rare sites of involvement. It is more common in women with dark skin. A differential diagnosis should be made in ulceration due to stasis dermatitis or skin involvement of tuberculosis.[5,6,8-10]

Hypopigmented Sarcoidosis

It is in the form of hypopigmented macules, papules, or nodules in patients with dark skin tones. Vitiligo, seborrheic dermatitis, leprosy, and pityriasis alba should be considered in the differential diagnosis.[5,6,8-10]

Ichthyosiform Sarcoidosis

They are sensitive and non-pruritic lesions in the form of gray, brown, scaly, and hyperpigmented plaques on the lower extremities. It should be differentiated from eczema.[5,6,8-10]

Lupus Pernio

They are hard, red-to-purple lesions consisting of papules or plaques. Lupus erythematosus, rhinophyma, lymphomas, cutaneous angiosarcoma, lupus vulgaris, and leprosy should be considered in the differential diagnosis. Lesions are seen on the nose, cheeks, lips, forehead, and ears. Bone involvement of the hands and feet is common in lupus pernio.[5,6,8,9] It is associated with sarcoidosis cases with upper respiratory tract involvement, and intrathoracic, ocular, and reticuloendothelial system involvement is more common.[10]

Erythema Nodosum

It is frequently found in patients of European origin. It is believed that ongoing inflammation is the cause. It is a nonspecific cutaneous finding frequently found in sarcoidosis patients, often on the anterior surface of the lower extremities. Tender, erythematous nodules that frequently accompany arthritis are its defining features. No granuloma is detected in biopsies. It undergoes spontaneous regression in six to eight weeks. Infection, nodular vasculitis, and thrombophlebitis should be considered in the differential diagnosis.[5-8] In studies, sarcoidosis was diagnosed in 10-22% of patients with erythema nodosum.[9,10]

Löfgren's Syndrome

It is a syndrome characterized by hilar lymphadenopathy, symmetrical polyarthralgia, anterior uveitis, and fever. It has a good prognosis and should be differentiated from infection-related conditions such as fungus or tuberculosis.[5,6,8-10]

Lichenoid Sarcoidosis

It is more common in children and accounts for 1-2% of cutaneous sarcoidosis. Multiple 1-3 mm-diameter flat or dome-shaped erythematous or skin-colored erythematous or skin-colored lesions are widely localized on the face, body, and extremities.[11]

Psoriasiform sarcoidosis

The sharply circumscribed squamous plaques seen in 0.9% of sarcoidosis patients are difficult to differentiate from psoriasis. In psoriasis, the distinction can be made by the scarless healing of the lesions.[12]

Verrucous Sarcoidosis

It is characterized by sharply circumscribed hyperkeratotic papillomatous lesions localized on the lower extremities. The warts may resemble Keratoacanthoma prurigo nodularis.[13]

Erythrodermic Sarcoidosis

It is a fusion of erythematous plaques, leaving intact areas in between. A biopsy is necessary to investigate the common causes of erythroderma.[14]

Necrobiosis-lipoidica-like Lesions

It is characterized by depressed pink-purplish plaques in the lower extremities. Sarcoidosis should be investigated in patients with necrobiosis lipoidica without diabetes.[15]

Livedo

It is a form characterized by erythematous-violaceous livedoid macules. Eye and central nervous system involvement is more common in these patients.[16]

Apart from all these cutaneous findings, there are some rare involvements. Alopecia, with or without scalp scarring, can be seen. Plaques, nodules, edema, and papules can be seen in the oral cheek mucosa, gingiva, hard palate, tongue, and posterior pharynx. Nail involvement is a rare cutaneous lesion, and genital sarcoidosis has been reported in case reports.[9-11]

TREATMENT

Topical or systemic corticosteroids, topical clobetasol and triamcinolone injections, chloroquine, and hydroxychloroquines are used in sarcoidosis. Methotrexate and cyclosporine are available for immunosuppressive treatment. In recent years, monoclonal antibodies, thalidomide, and

isotretinoin can be used as additional treatments. Drug reactions should be considered. Systemic immunosuppressives should be considered early in lupus pernio.[8-17]

CONCLUSION

Sarcoidosis is a multisystemic inflammatory disease of unknown etiology, affecting all organs and systems. While skin involvement in sarcoidosis is observed in one-third of cases, it may pose a diagnostic challenge due to its different morphology. The most common nonspecific lesions are erythema nodosum, calcifications, and prurigo. In general, early diagnosis is also beneficial.

ETHICAL DECLARATIONS

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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